

Inherited Metabolic Diseases (IMD) Program

List of Disorders, Covered Drugs, Supplements and Specialty Foods

Effective March 4, 2016

Ontario Public Drug Programs
Ministry of Health and Long-Term Care



Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

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Acronyms:

DIN = Drug Identification Number

PIN = Product Identification Number

NPN = Natural Product Number

SFS SKU = The Hospital for Sick Children's Specialty Food Shop stock number

SAP = Special Access Program

No DIN = Food Supplement or Chemical, no DIN assigned

HSC = Made at The Hospital for Sick Children, no commercial product available

Inquiries regarding the IMD Program should be directed to:

Ontario Public Drug Programs

5700 Yonge Street, 3rd Floor

Toronto ON M2M 4K5

Tel: 416-327-8109, Toll-free: 1-866-811-9893

Fax: 416-327-8123

Email: PublicDrugPrgrms.moh@ontario.ca

Inherited Metabolic Diseases (IMD) Program

List of Disorders, Covered Drugs, Supplements and Specialty Foods

About the IMD Program

Funding and administration of the Inherited Metabolic Diseases (IMD) program was transitioned to Ontario Public Drug Programs (OPDP) in February 2008. One goal of transitioning the IMD program to OPDP is to align it with other drug programs in Ontario. It also creates one point of access in the Ministry of Health and Long-Term Care (Ministry) for all drugs, with clear, consistent and evidence-based methods to make funding decisions.

The IMD subcommittee will evaluate requests for the addition of products and metabolic disorders to the IMD program's List of Disorders, Covered Drugs, Supplements and Specialty Foods (IMD program list). Membership of the subcommittee consists of metabolic and genetic specialists from each of the Ontario Newborn Screening Program regional treatment centres and the University Health Network, a pharmacist and a dietitian. The subcommittee will then make funding recommendations to the Executive Officer (EO), OPDP. For some drug products, the EO may also ask the Committee to Evaluate Drugs to provide a separate review and recommendation. The EO will make the final funding decision based on careful consideration of the subcommittee's recommendations, the public interest and sustainability of the Ontario Public Drug Program.

For new products to be listed under the Drugs and Supplements category on the IMD program list, requests must be submitted by a physician licensed to practice in Ontario. Completed applications will be assigned to a primary reviewer who will then submit a report to the IMD subcommittee. Both will be considered by the IMD subcommittee and their recommendations will then be forwarded to the Ministry and the EO.

For drug products approved for marketing by Health Canada [i.e., those with a drug identification number (DIN) and issued a Notice of Compliance (NOC)], it is the drug manufacturer's responsibility to follow the submission process used for products under consideration for listing on the Ontario Drug Benefit Formulary, and the requirements set out in the *Ontario Guidelines for Drug Submission & Evaluation*. This may include a submission to the Common Drug Review.

For drug products currently listed on the IMD program list, the Ministry has been working with the IMD subcommittee to specify, where appropriate, brand names, manufacturers, dosage forms and strengths, DINs, etc.

Provisional Listings

The IMD subcommittee has recommended an "approve with provisions" category for products and disorders discussed for consideration of inclusion on the IMD product list. Products and disorders with provisional approvals will require further review to determine their final status. Provisional listings are identified in italics.

Provisional listings for individual patients may not be reflected in the published version of the IMD program list. The requesting physician(s) will be notified by the Ministry of the final funding decision and the details of any listing provisions as recommended by the IMD subcommittee.

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Notable Changes for this Edition

New products and changes are **highlighted**.

Disorders

- 1 new disorder has been added: Glucose transporter 1 (GLUT1) deficiency syndrome (GLUT1-DS).

Drugs and Supplements

- Approved disorder(s) have been added to 4 drugs/ supplements.
- DIN/PIN/etc. has been clarified for **5-hydroxytryptophan**. Going forward, only 5-hydroxytryptophan products with valid Natural Product Number (NPN) or compounded from powder available in Canada will be covered.
- Alkaptonuria is removed as a funded disorder for nitisinone due to the lack of published evidence to support efficacy use in any cohort of asymptomatic or symptomatic alkaptonuria patients. The ministry also noted that there are no patients receiving nitisinone for this indication through the IMD Program. Funding remains in place for tyrosinemia type 1.

Food products and other supplements

- 2 new medical foods for tyrosinemia have been added: Tylactin RTD 15 Original and Tylactin Restore 10 Citrus
 - The Ministry has approved the addition of these Tylactin products noting the limited number of products available on the IMD list for patients with tyrosinemia, and the neutral cost to the IMD program.
 - The Ministry has also reviewed the Glytactin series of products for phenylketonuria (PKU) and has decided to hold making a decision on coverage of these products at this time. There is significantly more choice or products for PKU patients on the current IMD list compared to tyrosinemia. In addition, the proposed price of some Glytactin products exceeds the current price of available alternatives for PKU.
- Numerous “housekeeping” changes to update product SKUs or to remove discontinued products. Discontinued products (removed from list):
 - Phlexy-10 Capsule PKU Lophlex (46995)
 - PhenylAde PheBloc LNAA Tablets (SFS1489)

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Disorders Covered by the IMD Program

| CATEGORY | DISORDER |
|--|---|
| Amino Acid Disorders | 3-phosphoglycerate dehydrogenase deficiency |
| | Alkaptonuria |
| | Gyrate atrophy (ornithine aminotransferase deficiency) |
| | Homocystinuria (cystathionine beta-synthase [CBS] deficiency) |
| | Hyperphenylalaninemia, including phenylketonuria (PKU) |
| | Maple syrup urine disease (MSUD) (branched chain ketoacid dehydrogenase [BCKAD] deficiency) |
| | Tyrosinemia (includes Types I ,II & III) |
| | |
| Carbohydrate Disorders | Congenital disorder of glycosylation type 1b (CDG-1b) |
| | Congenital sucrase-isomaltase deficiency (CSID) (intestinal disaccharidase deficiency) |
| | Galactosemia |
| | Glycogen storage disorders (GSD) |
| | |
| Cholesterol Biosynthesis Disorders | Smith-Lemli-Opitz syndrome (SLOS) |
| | |
| Fatty Acid Oxidation Defects and Fat Metabolism Disorders | Abetalipoproteinemia |
| | Apoprotein C-II deficiency |
| | Carnitine palmitoyl transferase I (CPT I) deficiency |
| | Carnitine palmitoyl transferase II (CPT II) deficiency |
| | Carnitine acylcarnitine translocase (CACT) deficiency |
| | Carnitine uptake defect / carnitine transport defect / primary carnitine deficiency |
| | Chylomicron retention disease |
| | Glutaric aciduria Type II (GA II) |

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| CATEGORY | DISORDER |
|--|--|
| Fatty Acid Oxidation Defects and Fat Metabolism Disorders (cont'd) | Homozygous hypobetalipoproteinemia |
| | Lipoprotein lipase (LPL) deficiency |
| | Long chain 3-hydroxyacyl CoA dehydrogenase (LCHAD)/ Trifunctional protein deficiency |
| | Medium chain acyl CoA dehydrogenase (MCAD) deficiency |
| | Medium/short chain hydroxyacyl CoA dehydrogenase (M/SCHAD) deficiency |
| | Short chain acyl CoA dehydrogenase (SCAD) deficiency |
| | Very long chain 3-hydroxyacyl CoA dehydrogenase (VLCAD) deficiency |
| | |
| Lactic Acidosis (gluconeogenesis disorders) | Fructose-1,6-bisphosphatase deficiency |
| | Phosphoenol pyruvate carboxykinase (PEPCK) deficiency |
| | Pyruvate carboxylase (PC) deficiency |
| | Pyruvate dehydrogenase (PDH) deficiency |
| | |
| Mitochondrial Disorders | Primary mitochondrial disorder not otherwise specified |
| | Coenzyme Q10 deficiency |
| | Complex 1 deficiency |
| | Friedreich's ataxia |
| | Leber's hereditary optic neuropathy (LHON) |
| | Leigh's disease |
| | Mitochondrial encephalopathy, Lactic acidosis, Stroke-like episodes (MELAS) syndrome |
| | Mitochondrial myopathy |
| | |
| Organic Acid Disorders | 2-methylbutyryl-CoA dehydrogenase deficiency |

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| CATEGORY | DISORDER |
|--|--|
| Organic Acid Disorders (cont'd) | 2-methyl-3-hydroxybutyryl-CoA dehydrogenase (MHBD) deficiency / short branched chain hydroxyacyl CoA dehydrogenase (SBCHAD) deficiency |
| | 3-hydroxyisobutyric aciduria |
| | 3-hydroxy-3-methyl-glutaric (HMG) CoA lyase deficiency |
| | 3-methylcrotonyl-CoA carboxylase deficiency (3MCC) |
| | 3-methylglutaconic aciduria (includes Barth syndrome) |
| | β-Ketothiolase deficiency |
| | Glutaric aciduria Type I (GA I) |
| | HMG CoA lyase deficiency |
| | Isobutyryl-CoA dehydrogenase deficiency |
| | Isovaleric acidemia |
| | Malonic aciduria |
| | Methylmalonic acidemia (MMA) |
| | Propionic acidemia |
| Pyridoxine (B6)-dependent seizures (alpha-amino adipic semialdehyde dehydrogenase deficiency) [also listed under Vitamin/Cofactor Disorders] | |
| Urea Cycle Disorders | Argininemia |
| | Arginosuccinic acid lyase (AL or ASL) deficiency/ argininosuccinic aciduria (ASAuria) |
| | Carbamyl phosphate synthase (CPS) deficiency |
| | Citrullinemia [arginosuccinic acid synthetase (AS or ASS)] deficiency |
| | Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) |
| | N-acetyl glutamate synthetase (NAGS) deficiency |
| | Ornithine transcarbamylase (OTC) deficiency |

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| CATEGORY | DISORDER |
|-----------------------------------|---|
| Vitamin/Cofactor Disorders | Biopterin deficiency (includes dihydropteridine reductase [DHPR] deficiency) |
| | Cobalamin (B12) defect (includes Cbl C,D,G) |
| | Cobalamin (B12) transport deficiency, includes transcobalamin II (TC II) deficiency |
| | Hereditary deficiency of tocopherol transport protein |
| | Methylene tetrahydrofolate reductase (MTHFR) deficiency |
| | Multiple carboxylase deficiency (MCD) – Biotinidase deficiency |
| | Multiple carboxylase deficiency (MCD) – Holocarboxylase synthetase deficiency |
| | Pyridoxal-5-phosphate dependent epilepsy |
| | Pyridox(amine) 50-phosphate oxidase (PNPO) deficiency |
| | Pyridoxine (B6)-dependent seizures (alpha-aminoacidic semialdehyde dehydrogenase deficiency) [also listed under Organic Acid Disorders] |
| | Vitamin E (tocopherol) deficiency |
| | |
| Miscellaneous Disorders | Acute intermittent porphyria |
| | Cerebrotendinous xanthomatosis |
| | Creatine synthesis/transport deficiency |
| | Cystinosis |
| | Cystinuria |
| | Erythropoietic protoporphyria (EPP) |
| | Glucose transporter 1 (GLUT1) deficiency syndrome (GLUT1-DS) |
| | Hypercalcemia secondary to a listed disease (please specify) |
| | Lysinuric protein intolerance (LPI) |
| | Menkes disease |
| | Renal tubular acidosis (RTA) secondary to a listed disease (please specify on registration form) |

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Drugs and Supplements

Note: The Ministry continues to receive requests to substitute alternate brands of drugs when the listed brands or DINs have been discontinued. In addition, the transition of many over-the-counter products to natural health products under Health Canada's Natural Health Products Regulations has resulted in many DINs being changed to a **Natural Product Number (NPN)**. The Ministry will consider reasonable substitutions if there is little or no difference in cost. Treatment centre pharmacies can contact OPDP with any questions regarding coverage and billing of Drugs and Supplements under the IMD program.

A product in *italics* is considered a provisional listing pending further review by the MOHLTC. Depending on the outcome of this review, the MOHLTC may decide not to fund the product(s) or the listed indication(s) in the future.

| DRUGS AND SUPPLEMENTS | | |
|--------------------------------|--|---|
| Drug/Supplement | Approved Disorder(s) | DIN/PIN/ETC. |
| 5-hydroxytryptophan | Biopterin deficiency; Pyridoxine-dependent epilepsy (PDE) | Product must have a valid NPN or may be compounded from powder |
| Ammonul | Urea Cycle Disorders | SAP |
| Beta-Carotene (Lumitene, oral) | Erythropoietic Protoporphyrin (EPP) | |
| Betaine (Cystadane) | Homocystinuria (includes homocystinuria secondary to a listed disorder) | SAP |
| Biotin | Biotinidase deficiency; Holocarboxylase synthetase deficiency; Propionic acidemia; Pyruvate carboxylase deficiency | No DIN |
| <i>Chenodeoxycholic acid</i> | <i>Cerebrotendinous Xanthomatosis</i> | <i>No DIN</i> |
| <i>Cholesterol</i> | <i>Smith-Lemli-Opitz syndrome (SLOS)</i> | <i>No DIN</i> |
| Citrate C | Renal tubular acidosis, secondary to another listed disorder | HSC |
| Citrulline | Urea cycle disorders; Lysinuric protein intolerance | No DIN |
| <i>Coenzyme-Q</i> | <i>Mitochondrial disorders; Glutaric Aciduria II, secondary mitochondrial defects</i> | <i>No DIN</i> |

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| DRUGS AND SUPPLEMENTS | | |
|--|---|--------------|
| Drug/Supplement | Approved Disorder(s) | DIN/PIN/ETC. |
| Copper histidine | Menkes disease | HSC |
| <i>Creatine</i> | <i>Mitochondrial myopathies; Creatine synthesis/transport deficiency; Gyrate atrophy</i> | SAP |
| <i>Cupric Chloride</i> | <i>Menkes disease</i> | |
| Cysteamine (Cystagon or cysteamine powder for compounded products) | Cystinosis | SAP |
| Dichloroacetate | Pyruvate dehydrogenase deficiency | SAP |
| Glycine | Isovaleric academia; Creatine transport deficiency | No DIN |
| Hemin (Normosang) | Acute intermittent porphyria Note: Eligibility criteria for IMD program funding (all criteria must be met): <ol style="list-style-type: none"> 1. <i>Diagnosis of acute intermittent porphyria should be confirmed by biochemical and molecular testing, and;</i> 2. <i>Hemin (Normosang) infusions should only be given under supervision and be prescribed by hematologists, gastroenterologists or metabolic specialists, and;</i> 3. <i>For continued funding, yearly follow up report should be submitted to the IMD program by the prescribing physician.</i> | SAP |
| Hydroxycobalamin | Cobalamin defect; Methylmalonic acidemia | SAP |
| L-arginine | Lysinuric protein intolerance; MELAS syndrome; Pyridoxine (B6)-dependent seizures; Urea cycle disorders; Creatine transport deficiency | No DIN |
| L-aspartic acid | Pyruvate carboxylase deficiency | No DIN |

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| DRUGS AND SUPPLEMENTS | | |
|--------------------------------------|---|--|
| Drug/Supplement | Approved Disorder(s) | DIN/PIN/ETC. |
| L-carnitine | Organic acidemias; Fatty acid oxidation defects; carnitine uptake defect, carnitine deficiency secondary to another listed disorder | DIN=02144336, DIN=02144328, DIN=02144344 |
| L-isoleucine | MSUD, organic acid disorders | No DIN |
| L-lysine | Lysinuric protein intolerance | No DIN |
| L-valine | MSUD, organic acid disorders | No DIN |
| <i>Lipoic Acid</i> | <i>Mitochondrial disorders</i> | |
| Mannose | Congenital Disorders of Glycosylation | No DIN |
| <i>Niacinamide</i> | <i>Mitochondrial cytopathies</i> | <i>No DIN</i> |
| Nitisinone (NTBC, Orfadin) | Tyrosinemia Type I | SAP |
| Ornithine HCl, ornithine L-aspartate | Urea cycle disorders; HHH; Creatine synthesis/transport deficiency | No DIN |
| Phosphocysteamine | Cystinosis | SAP |
| Pyridoxal-5-phosphate | Pyridoxal-5-phosphate dependent epilepsy; Pyridox(amine) 50-phosphate oxidase (PNPO) deficiency | Product must have a valid NPN |
| Pyridoxine HCl | Homocystinuria (includes homocystinuria secondary to a listed disorder); Pyridoxine (B6)-dependent seizures | DIN 00497517 NPN 00232475 NPN 00268607 |
| <i>Riboflavin</i> | <i>Mitochondrial disorders; Glutaric aciduria</i> | <i>No DIN</i> |
| Sapropterin (Kuvan) ¹ | Biopterin deficiency | DIN 02350580 |
| Serine | 3-phosphoglycerate dehydrogenase deficiency | No DIN |
| Sodium benzoate | Creatine synthesis/transport deficiency; Urea cycle disorders | No DIN |

¹ Sapropterin (Kuvan[®]) is not reimbursed under the IMD program for the treatment of phenylketonuria (PKU). Funding for PKU is considered under the Exceptional Access Program (EAP) only. Under an agreement with the manufacturer, the maximum reimbursable price for Kuvan under the IMD program is \$33.00 per 100mg tablet.

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| DRUGS AND SUPPLEMENTS | | |
|---|--|---|
| Drug/Supplement | Approved Disorder(s) | DIN/PIN/ETC. |
| <i>Sodium phenylbutyrate</i> ² | <i>Urea cycle disorders</i> | |
| <i>Thiamine (Vitamin B1)</i> | <i>Mitochondrial cytopathies; Thiamine deficiency in the presence of IMD</i> | <i>DIN 00816078 NPN 00268631 NPN 00294853</i> |
| <i>Tiopronin (Thiola)</i> | <i>Cystinuria</i> | <i>SAP</i> |
| <i>Vitamin A/D/E/K</i> | <i>Abetalipoproteinemia; Mitochondrial disorders</i> | Product must have a valid DIN or NPN |
| <i>Vitamin A</i> | <i>Abetalipoproteinemia</i> | Product must have a valid DIN or NPN |
| <i>Vitamin B50 Complex</i> | <i>Mitochondrial disorders (for patients requiring multiple B vitamins and where the cost of vitamin B50 complex does not exceed the cost of the individual vitamins).</i> | Product must have a valid NPN |
| <i>Vitamin D</i> | <i>Abetalipoproteinemia</i> | Product must have a valid DIN or NPN |
| <i>Vitamin E</i> | <i>Abetalipoproteinemia; Hereditary deficiency of tocopherol transport protein; Mitochondrial disorders; Vitamin E deficiency</i> | Product must have a valid DIN or NPN |
| <i>Vitamin K1</i> | <i>Mitochondrial disorders</i> | Product must have a valid DIN or NPN |
| <i>Vitamin K3</i> | <i>Mitochondrial disorders</i> | Product must have a valid DIN or NPN |

² The Ministry of Health and Long-Term Care is aware that the Buphenyl formulation of sodium phenylbutyrate may no longer be available in Canada. The IMD program will provide funding for Pheburane[®] on an interim basis pending a further review to determine the final listing status.

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Medical Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|--|---|------------------|-----------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| BCAD 1 | MSUD | Mead Johnson | SFS1071 |
| Camino pro MSUD Drink, Fruit Punch | MSUD | Cambrooke Foods | 61002 |
| Camino pro MSUD Drink, Pina Colada | MSUD | Cambrooke Foods | 61001 |
| Camino pro PKU Drink, Fruit Punch | PKU, Biopterin deficiency | Cambrooke Foods | 31002 |
| Camino pro PKU Drink, Pina Colada | PKU, Biopterin deficiency | Cambrooke Foods | 31001 |
| Complex MSUD Amino Acid Bars | MSUD | Nutricia | 47021 |
| Complex MSUD Amino Acid Blend | MSUD | Nutricia | 47031 |
| Complex MSUD Vanilla Flavoured Drink Mix | MSUD | Nutricia | 47024 |
| Complex Essential MSD Drink Mix – Vanilla | MSUD | Nutricia | 59720 |
| Complex MSUD Amino Acid Blend | MSUD | Nutricia | 47031 |
| Cyclinex–1 | Urea cycle disorders, Creatine synthesis/transport deficiency | Abbott Nutrition | 47012 |
| Cyclinex–2 | Urea cycle disorders, Creatine synthesis/transport deficiency | Abbott Nutrition | 47013 |
| Essential Amino Acid Mix | Urea cycle disorders, Gyrate Atrophy, Creatine synthesis/transport deficiency | Nutricia | SFS1556 |
| EAA Supplement | Urea cycle disorders, Gyrate Atrophy, Creatine synthesis/transport deficiency | Vitaflo | 47052 |

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| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|---|------------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| GA1 Express 15 | Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures | Vitaflo | SFS1075 |
| GA1 Gel, Unflavoured | Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures | Vitaflo | 18888 |
| GlutarAde GA-1 Amino Acid Blend | Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures | Nutricia | SFS1074 |
| Glutarade Essential GA-1 Drink Mix | Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures | Nutricia | SFS1310 |
| Glutarex–1 | Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures | Abbott Nutrition | 46998 |
| Glutarex–2 | Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures | Abbott Nutrition | 46999 |
| HCU Cooler 10 red | Homocystinuria | Vitaflo | SFS1076 |
| HCU Cooler 15 orange | Homocystinuria | Vitaflo | 18889 |
| HCU Cooler 15 red | Homocystinuria | Vitaflo | SFS0538 |
| HCU Cooler 20 red | Homocystinuria | Vitaflo | SFS1077 |
| HCU express | Homocystinuria | Vitaflo | 1809 |
| HCU Express 20 | Homocystinuria | Vitaflo | SFS1078 |
| HCU gel | Homocystinuria | Vitaflo | 1808 |
| HCU Lophlex LQ Mixed Berry Blast | Homocystinuria | Nutricia | SFS1086 |
| Hominex – 1 | Homocystinuria | Abbott Nutrition | 46986 |
| Hominex – 2 | Homocystinuria | Abbott Nutrition | 46979 |
| IVA Cooler 15 red | Isovaleric acidemia | Vitaflo | SFS1315 |
| Ketonex – 1 | MSUD | Abbott Nutrition | 47007 |

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| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|--|--|------------------|-----------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| Ketonex – 2 | MSUD | Abbott Nutrition | 47009 |
| MMA/PA Cooler 15 red | Methylmalonic Acidemia, Propionic Acidemia | Vitaflo | SFS1314 |
| MMA/PA express | Methylmalonic Acidemia, Propionic Acidemia | Vitaflo | 47054 |
| MMA/PA gel | Methylmalonic Acidemia, Propionic Acidemia | Vitaflo | 47053 |
| MSUD Analog | MSUD | Nutricia | 47035 |
| MSUD Cooler 10 red | MSUD | Vitaflo | SFS1079 |
| MSUD Cooler 15 orange | MSUD | Vitaflo | 1822 |
| MSUD Cooler 15 red | MSUD | Vitaflo | SFS0536 |
| MSUD Cooler 20 red | MSUD | Vitaflo | SFS1080 |
| MSUD Express (unflavoured sachet) | MSUD | Vitaflo | 1807 |
| MSUD Express 20 | MSUD | Vitaflo | SFS1081 |
| MSUD Gel | MSUD | Vitaflo | 1806 |
| MSUD Lophlex LQ Mixed Berry Blast | MSUD | Nutricia | SFS1087 |
| MSUD Maxamaid | MSUD | Nutricia | 78964 |
| MSUD Maxamum | MSUD | Nutricia | 47022 |
| Periflex Advance Orange | PKU, Biopterin deficiency | Nutricia | 47065 |
| Periflex Advance Unflavoured | PKU, Biopterin deficiency | Nutricia | 47064 |
| Periflex Infant | PKU, Biopterin deficiency | Nutricia | 11400 |
| Periflex Junior Orange | PKU, Biopterin deficiency | Nutricia | 47062 |

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|---|---------------------------|------------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| PKU Periflex Junior Plus, Orange | PKU, Biopterin deficiency | Nutricia | SFS1805 |
| Periflex Junior Plus Plain | PKU, Biopterin deficiency | Nutricia | SFS1488 |
| Periflex LQ Berry | PKU, Biopterin deficiency | Nutricia | 1256 |
| Periflex LQ Orange | PKU, Biopterin deficiency | Nutricia | 1255 |
| Phenex–1 | PKU, Biopterin deficiency | Abbott Nutrition | 47005 |
| Phenex–2 | PKU, Biopterin deficiency | Abbott Nutrition | 47003 |
| Phenex–2 Vanilla | PKU, Biopterin deficiency | Abbott Nutrition | 47028 |
| PhenylAde 40 Citrus Flavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 1319 |
| Phenylade 40 Unflavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 1320 |
| PhenylAde 60 – Unflavoured Drink Mix (can) | PKU, Biopterin deficiency | Nutricia | SFS0454 |
| PhenylAde 60 – Unflavoured Drink Mix (pouch) | PKU, Biopterin deficiency | Nutricia | SFS0531 |
| PhenylAde 60 – Vanilla Flavoured Drink Mix (can) | PKU, Biopterin deficiency | Nutricia | 47088 |
| PhenylAde 60 – Vanilla Flavoured Drink Mix (pouch) | PKU, Biopterin deficiency | Nutricia | SFS0532 |
| PhenylAde Chocolate Flavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 47044 |

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| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|---------------------------|------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| PhenylAde Orange Flavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 47023 |
| PhenylAde Strawberry Flavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 47025 |
| Phenylade Vanilla Flavoured Drink Mix | PKU, Biopterin deficiency | Nutricia | 47017 |
| PhenylAde Essential Drink Mix- Chocolate (can) | PKU, Biopterin deficiency | Nutricia | 9501 |
| PhenylAde Essential Drink Mix- Orange (can) | PKU, Biopterin deficiency | Nutricia | 9503 |
| PhenylAde Essential Drink Mix- Strawberry (can) | PKU, Biopterin deficiency | Nutricia | 9504 |
| PhenylAde Essential Drink Mix- Vanilla (can) | PKU, Biopterin deficiency | Nutricia | 9502 |
| PhenylAde Essential Drink Mix- Chocolate (pouch) | PKU, Biopterin deficiency | Nutricia | 95014 |
| PhenylAde Essential Drink Mix- Orange (pouch) | PKU, Biopterin deficiency | Nutricia | 95034 |
| PhenylAde Essential Drink Mix- Strawberry (pouch) | PKU, Biopterin deficiency | Nutricia | 95044 |
| PhenylAde Essential Drink Mix- Vanilla (pouch) | PKU, Biopterin deficiency | Nutricia | 95024 |
| PhenylAde Amino Acid Bars Chocolate Flavoured | PKU, Biopterin deficiency | Nutricia | 470181 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|---------------------------|------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| PhenylAde Amino Acid Bars Chocolate Flavoured | PKU, Biopterin deficiency | Nutricia | 470182 |
| PhenylAde Amino Acid Bars White Chocolate Flavoured | PKU, Biopterin deficiency | Nutricia | 47019 |
| PhenylAde MTE Amino Acid Blend Unflavoured | PKU, Biopterin deficiency | Nutricia | 47015 |
| PhenylAde MTE Amino Acid Blend (pouch) | PKU, Biopterin deficiency | Nutricia | 1315 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|---|------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| PhenylAde PheBloc LNAA Pouches | <p>PKU</p> <p>Note: Eligibility criteria for IMD program funding (all criteria must be met):</p> <ol style="list-style-type: none"> 1. <i>The patient will have untreated/late-diagnosed PKU <u>OR</u> early diagnosed PKU with elevated blood phenylalanine concentrations beyond his/her ability to correct with the phenylalanine-restricted diet;<u>AND</u></i> 2. <i>The patient will have blood phenylalanine concentrations which are chronically above his/her treatment goal;<u>AND</u></i> 3. <i>The patient will be 13 years of age or over;<u>AND</u></i> 4. <i>The patient will not be pregnant or planning a pregnancy;<u>AND</u></i> 5. <i>The patient will not also be treated with sapropterin dihydrochloride (Kuvan);<u>AND</u></i> 6. <i>It is recommended that the patient use PheBloc (divided evenly between meals) to provide between 0.25-0.5 grams protein equivalent/kg/day (use adjusted body weight for overweight & obese individuals);<u>AND</u></i> 7. <i>It is recommended that the patient supplement the intake of protein from PheBloc with natural protein foods (and a “medical food” if necessary), to achieve a total protein intake of no less than 1 gram protein/kg/day (use adjusted body weight for overweight & obese individuals); <u>AND</u></i> 8. <i>The patient will use PheBloc only under the direction of a metabolic geneticist/metabolic dietitian.</i> <p><i>Continued use of PheBloc is approved only for patients who are assessed by the metabolic geneticist to have had an improvement in signs and/or symptoms associated with elevated blood phenylalanine concentrations.</i></p> | Nutricia | SFS1487 (pouches), |

Inherited Metabolic Diseases (IMD) Program
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| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|---------------------------|--------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| Phenyl-Free 1 | PKU, Biopterin deficiency | Mead Johnson | 46988 |
| Phenyl-Free 2 | PKU, Biopterin deficiency | Mead Johnson | 47029 |
| Phenyl-Free 2HP | PKU, Biopterin deficiency | Mead Johnson | 47030 |
| Phlexy-10 Drink Mix – Blackcurrant/Apple | PKU, Biopterin deficiency | Nutricia | 46996 |
| Phlexy-10 Drink Mix – Tropical Surprise | PKU, Biopterin deficiency | Nutricia | 1231 |
| PKU Air 15 green | PKU, Biopterin deficiency | Vitaflo | SFS1311 |
| PKU Air 20 gold | PKU, Biopterin deficiency | Vitaflo | SFS1313 |
| PKU Air 20 green | PKU, Biopterin deficiency | Vitaflo | SFS1312 |
| PKU Cooler 10 orange | PKU, Biopterin deficiency | Vitaflo | 47055 |
| PKU Cooler 10 purple | PKU, Biopterin deficiency | Vitaflo | 47056 |
| PKU Cooler 10 red | PKU, Biopterin deficiency | Vitaflo | SFS0533 |
| PKU Cooler 15 orange | PKU, Biopterin deficiency | Vitaflo | 47049 |
| PKU Cooler 15 purple | PKU, Biopterin deficiency | Vitaflo | 47051 |
| PKU Cooler 15 red | PKU, Biopterin deficiency | Vitaflo | SFS0534 |
| PKU Cooler 20 orange | PKU, Biopterin deficiency | Vitaflo | 47057 |
| PKU Cooler 20 purple | PKU, Biopterin deficiency | Vitaflo | 47058 |
| PKU Cooler 20 red | PKU, Biopterin deficiency | Vitaflo | SFS0535 |
| PKU Cooler 10 – White | PKU, Biopterin deficiency | Vitaflo | 98745 |
| PKU Cooler 15 – White | PKU, Biopterin deficiency | Vitaflo | 98756 |
| PKU Cooler 20 – White | PKU, Biopterin deficiency | Vitaflo | 98766 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|--|------------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| PKU Express lemon | PKU, Biopterin deficiency | Vitaflo | 1805 |
| PKU Express orange | PKU, Biopterin deficiency | Vitaflo | 1804 |
| PKU Express tropical | PKU, Biopterin deficiency | Vitaflo | 1818 |
| PKU Express unflavoured | PKU, Biopterin deficiency | Vitaflo | 1803 |
| PKU Express 20 lemon | PKU, Biopterin deficiency | Vitaflo | SFS1820 |
| PKU Express 20 orange | PKU, Biopterin deficiency | Vitaflo | SFS1821 |
| PKU Express 20 tropical | PKU, Biopterin deficiency | Vitaflo | SFS1822 |
| PKU Express 20 unflavoured | PKU, Biopterin deficiency | Vitaflo | SFS1082 |
| PKU gel orange | PKU, Biopterin deficiency | Vitaflo | 1801 |
| PKU gel raspberry | PKU, Biopterin deficiency | Vitaflo | 47045 |
| PKU gel unflavoured | PKU, Biopterin deficiency | Vitaflo | 1800 |
| PKU Lophlex Berry | PKU, Biopterin deficiency | Nutricia | 1248 |
| PKU Lophlex LQ Juicy Orange | PKU, Biopterin deficiency | Nutricia | SFS0835 |
| PKU Lophlex LQ Mixed Berry Blast | PKU, Biopterin deficiency | Nutricia | SFS0834 |
| PKU Lophlex LQ Tropical | PKU, Biopterin deficiency | Nutricia | SFS0452 |
| PKU Lophlex Orange | PKU, Biopterin deficiency | Nutricia | 47000 |
| Propimex-1 | Methylmalonic acidemia, Propionic acidemia | Abbott Nutrition | 47008 |
| Propimex-2 | Methylmalonic acidemia, Propionic acidemia | Abbott Nutrition | 47010 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|---|--|------------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| Tylactin RTD 15, Original | Tyrosinemia | Cambrooke | SFS1804 |
| Tylactin Restore 10, Citrus | Tyrosinemia | Cambrooke | SFS1803 |
| Tyrex-1 | Tyrosinemia | Abbott Nutrition | 47006 |
| Tyrex-2 | Tyrosinemia | Abbott Nutrition | 47016 |
| TYROS 1 | Tyrosinemia | Mead Johnson | SFS1072 |
| TYR Cooler 10 red | Tyrosinemia | Vitaflo | SFS1083 |
| TYR Cooler 15 orange | Tyrosinemia | Vitaflo | 67896 |
| TYR Cooler 15 red | Tyrosinemia | Vitaflo | SFS0537 |
| TYR Cooler 20 red | Tyrosinemia | Vitaflo | SFS1084 |
| TYR Express | Tyrosinemia | Vitaflo | 1811 |
| TYR Express 20 | Tyrosinemia | Vitaflo | SFS1085 |
| TYR gel | Tyrosinemia | Vitaflo | 46899 |
| TYR Lophlex LQ Mixed Berry Blast | Tyrosinemia | Nutricia | SFS1088 |
| UCD Anamix Junior, unflavoured | Creatine synthesis/transport deficiency, HHH, Gyrate atrophy, Urea cycle disorders | Nutricia | SFS1089 |
| UCD Anamix Junior, vanilla | Creatine synthesis/transport deficiency, HHH, Gyrate atrophy, Urea cycle disorders | Nutricia | SFS1090 |
| I-Valex-1 | Isovaleric acidemia | Abbott Nutrition | 46993 |
| I-Valex-2 | Isovaleric acidemia | Abbott Nutrition | 46992 |
| XLeu Analog | Isovaleric acidemia | Nutricia | 47040 |
| XLeu Maxamaid | Isovaleric acidemia | Nutricia | 78965 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|--|---|------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| XLeu Maxamum | Isovaleric acidemia | Nutricia | 78966 |
| XLys, XTrp Analog | Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures | Nutricia | 47037 |
| XLys, XTrp Maxamaid | Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures | Nutricia | 47050 |
| XLys, XTrp Maxamum | Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures | Nutricia | 78967 |
| XMet Analog | Homocystinuria | Nutricia | 47036 |
| XMet Maxamaid | Homocystinuria | Nutricia | 78968 |
| XMet Maxamum | Homocystinuria | Nutricia | 46987 |
| XMTVI Analog | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia | 78969 |
| XMTVI Maxamaid | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia | 78970 |
| XMTVI Maxamum | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia | 78971 |
| XPhe Maxamaid Artificially Orange Flavoured Powder | PKU, Biopterin deficiency | Nutricia | 46985 |
| XPhe Maxamaid Unflavoured Powder | PKU, Biopterin deficiency | Nutricia | 46977 |
| XPhe Maxamum Artificially Orange Flavoured Powder (can) | PKU, Biopterin deficiency | Nutricia | 46984 |
| XPhe Maxamum Artificially Orange Flavoured Powder (sachet) | PKU, Biopterin deficiency | Nutricia | 1250 |
| XPhe Maxamum Unflavoured Powder (can) | PKU, Biopterin deficiency | Nutricia | 46989 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS) | | | |
|--|---------------------------|------------|-------------------------|
| Product | Disorder(s) | Mfr | PIN/ SFS SKU |
| XPhe Maxamum Unflavoured Powder (sachet) | PKU, Biopterin deficiency | Nutricia | 1251 |
| XPhe, XTyr Analog | Tyrosinemia | Nutricia | 47038 |
| XPhe, XTyr Maxamaid Artificially Orange Flavoured Powder | Tyrosinemia | Nutricia | 46990 |
| XPhen, XTyr Maxamum | Tyrosinemia | Nutricia | 1253 |
| XPTM Analog | Tyrosinemia | Nutricia | 47039 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

Special Low Protein Foods

| SPECIAL LOW PROTEIN FOODS | |
|---|--------------------|
| Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency | |
| Product | PIN/SFS SKU |
| Applied Nutrition Homestyle Blueberry Muffin Mix | 38126 |
| Applied Nutrition Cinnamon Chip Flavour Muffin Mix | 30400 |
| Applied Nutrition Homestyle Sugar Cookie Mix | 38127 |
| Applied Nutrition Homestyle Yellow Cake Mix | 38128 |
| Aproten Crispbread Rusks 260g | 31128 |
| Aproten Crackertoast 250g | 31130 |
| Aproten Farina/Pizza Mix | 31172 |
| Aproten Low protein Bread | 0255 |
| Aproten Pasta: ditalini 500g | 311702 |
| Aproten Pasta: fusilli 500g | 311703 |
| Aproten Pasta: spaghetti 500g | 311706 |
| Aproten Pasta: penne 500g | 311704 |
| Aproten Pasta: anellini 500g | 311701 |
| Aproten Pasta: rigatoni 500g | 311705 |
| Aproten Pasta: fettucine 250g | 311707 |
| Aproten Pasta: tagliatelle 250g | 311708 |
| Aproten Biscotto 180g | 31171 |
| Aproten Rice (Chicchi) 500g | 12163 |
| Cambrooke All-Purpose Baking Mix (2 lbs) | SFS1355 |
| Cambrooke Artisan Buns | 401010 |
| Cambrooke Bagels plain | SFS0444 |
| Cambrooke Brooklyn Dog Buns | 40616 |
| Cambrooke Focaccia Sticks - Italian (8 sticks) | 40112 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| SPECIAL LOW PROTEIN FOODS | |
|---|--------------------|
| Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency | |
| Product | PIN/SFS SKU |
| Cambrooke Foods – American Cheese Slices | 40311 |
| Cambrooke Foods – Camburger buns | 40124 |
| Cambrooke Foods – Camburgers | 2901 |
| Cambrooke Foods – Cheese Ravioli | 40413 |
| Cambrooke Foods – Pasta Elbows | 40455 |
| Cambrooke Foods – Pasta, Portabella Spinach Ravioli | 40116 |
| Cambrooke Foods – Swiss Cheese Slices | 40312 |
| Cambrooke Foods - Tweekz | 40902 |
| Cambrooke Foods –Tomato Tortilla Wraps | 2119 |
| Cambrooke Foods –Tortilla Wraps | 2118 |
| Cambrooke Homestyle White Bread (1 loaf) | 40110 |
| Cambrooke Imitation Mozzarella Shredded Cheese | 40315 |
| Cambrooke Macaroni & Cheese | 40406 |
| Cambrooke MixQuick Multi-Purpose Batter (2 lbs) | SFS 1354 |
| Cambrooke Pita Pockets | SFS0226 |
| Cambrooke Short Grain Japanese Rice (1kg) | 40407 |
| Cambrooke Tuscan Pizza Shells (4 shells) | 40115 |
| Cambrooke Veggie Meatballs | 40911 |
| Country Sunrise Chicken Flavoured Pattie/Nugget Mix | SFS0233 |
| Country Sunrise Imitation Peanut Butter and Honey Spread | 0165 |
| Country Sunrise Instant Mashed Potatoes | 0065 |
| Country Sunrise Mushroom Burger Mix | 0034 |
| Country Sunrise Scrambled Egg/Omelet Mix | 0105 |
| Country Sunrise Soft Tortillas | SFS0695 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| SPECIAL LOW PROTEIN FOODS | |
|---|-------------|
| Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency | |
| Product | PIN/SFS SKU |
| Country Sunrise Southwest Burger Mix | 0035 |
| Country Sunrise Vegetable Hot Dog Mix | 38346 |
| Dairy-Free milk packets | 1517 |
| D.S. (Dietary Specialties) Elbows 250g | 2293 |
| D.S. Spanish Rice | 1021 |
| D.S. OHZ in Sauce | 1022 |
| D.S. Imitation Peanut Butter 16oz | 2303 |
| D.S. Imitation Rice 500g | 2295 |
| D.S. Imitation Macaroni & Cheese 6oz | 2306 |
| D.S. Lasagna 100g | 2543 |
| D.S. Tri-coloured Shells 250g | 2290 |
| D.S. Porridge 500g | 2292 |
| D.S. Herb & Garlic Ziti 250g | 2291 |
| D.S. Bread Machine Baking Mix 1.8kg | 2294 |
| George Washington Seasoning: brown 31g | 141702 |
| George Washington Seasoning: golden 31g | 141701 |
| Homestyle Fudge Brownie Mix | SFS0230 |
| Juvela Low Protein Mix (Nutricia) | 77444 |
| Kingsmill Egg Replacer 350 g | 31197 |
| Kingsmill Vacuum Packed Unimix Bread 550g | 31219 |
| Kingsmill Unimix All Purpose Baking Mix | 31118 |
| La Tiara Taco Shells | SFS0079 |
| Loprofin Pasta: animal pasta 500g | 114155 |
| Loprofin Pasta: fusilli 500g | 126113 |

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| SPECIAL LOW PROTEIN FOODS | |
|---|--------------------|
| Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency | |
| Product | PIN/SFS SKU |
| Loprofin Pasta: lasagna (Nutricia) | SFS0515 |
| Loprofin Pasta: penne 500g | 126111 |
| Loprofin Pasta: spaghetti 500g | 126112 |
| Loprofin Pasta: vermicelli 250g | 12610 |
| Loprofin Crackers 150g | 12650 |
| Loprofin L/P Baking Mix 500g | 38125 |
| Loprofin Breakfast Cereal 375g | 12612 |
| Loprofin Chocolate Cake Mix | SFS0211 |
| Loprofin Rice | SFS0116 |
| Milupa Ip Bar | 30911 |
| Milupa Ip Drink | 12652 |
| Milupa Ip Dry Cereal Chocolate Ringlets | 30947 |
| Milupa Ip Dry Cereal Flakes | 30940 |
| Milupa Ip Fruity Cereal Mix – Apple Banana | 65991 |
| Milupa Ip Fruity Cereal Mix – Pear | 65992 |
| PKU Perspectives – Sandwichmate Imitation Cheese Slices | SFS0871 |
| PKU Perspectives – Shredmate Imitation Shredded Mozzarella Cheese | SFS1091 |
| Taste Connections – Low Protein Bread Mix | 450 |
| Taste Connections – Low Protein Multi-Baking Mix | 451 |
| Taste Connections – Low Protein Versa Mix | 4522 |
| Walden Farms Peanut Spread | SFS0080 |
| Wel-Plan Baking Mix 400g | 31235 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

Other Supplements

| OTHER SUPPLEMENTS | | |
|------------------------------------|---|-----------------|
| Product | Disorder(s) | PIN/ SFS SKU |
| Complete Amino Acid Mix (Nutricia) | Carbohydrate disorders, Lactic acidosis, where a modular approach is required | SFS1557 |
| Duocal | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 46982 |
| Flavour Pacs– Orange | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 1813 |
| Flavour Pacs – Lemon | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 1812 |
| Flavour Pacs – Raspberry | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 1814 |
| Flavour Pacs– Blackcurrant | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 1815 |
| Flavour Pacs- Tropical flavour | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 1819 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

| OTHER SUPPLEMENTS | | |
|--|---|-----------------|
| Product | Disorder(s) | PIN/ SFS SKU |
| <p>Glycosade (high-amylopectin-containing cornstarch)</p> | <p>Glycogen storage disease (GSD)</p> <p>Note: Eligibility criteria for IMD program funding (all criteria must be met:</p> <ol style="list-style-type: none"> 1. <i>The patient will have a diagnosis of glycogen storage disease type 1a/b or type 3 which requires cornstarch feedings overnight for metabolic control; AND</i> 2. <i>The patient will be ≥24 months of age; AND</i> 3. <i>The patient will use Glycosade, only under the direction of a metabolic geneticist/metabolic dietitian; AND</i> 4. <i>The patient will use Glycosade to maintain blood sugars overnight. Regular (i.e. unmodified) cornstarch will continue to be the standard treatment during the day.</i> <p>EXCEPTION: <i>If all attempts with conventional therapy (regular cornstarch & diet) during the day fail to achieve metabolic control in a patient, the metabolic geneticist may wish to consider a trial of Glycosade during the day.</i></p> <p><i>Continued use of Glycosade is approved only for patients who demonstrate improved metabolic control.</i></p> <p>Glucose transporter 1 deficiency syndrome (GLUT1-DS)* with the following eligibility criteria:</p> <ul style="list-style-type: none"> o <i>The patient has confirmed diagnosis of GLUT1-DS; and</i> o <i>The patient will use Glycosade to maintain blood sugars overnight along with low glycemic index diet during the day; and</i> <p>Patient has demonstrated failure to comply with ketogenic diet.</p> | <p>SFS0624</p> |
| <p>Liquigen</p> | <p>Abetalipoproteinemia, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic Acidosis, Mitochondrial disorders</p> | <p>SFS1490</p> |
| <p>MCT Pro-Cal</p> | <p>Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders</p> | <p>22990</p> |
| <p>MCT Oil</p> | <p>Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders</p> | <p>39409</p> |

Inherited Metabolic Diseases (IMD) Program
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| OTHER SUPPLEMENTS | | |
|---|---|-----------------|
| Product | Disorder(s) | PIN/ SFS SKU |
| Microlipid | Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders | 74410 |
| PFD 1 | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | SFS0539 |
| Phlexy-Vits | Amino acid disorders, Carbohydrate disorders, Fatty Acid Oxidation Defects, Lactic Acidosis, Mitochondrial Disorders, Organic acid disorders, Urea cycle disorders, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Amish Microcephaly | 46994 |
| Polycal | Abetalipoproteinemia, Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | SFS1654 |
| Pro-Phree | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 47011 |
| Protifar | Carbohydrate disorders | 79645 |
| ProViMin Powder | Abetalipoproteinemia, Carbohydrate disorders | 47026 |
| Quick Thick, LP/GF | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate Disorders, Mitochondrial disorders, Fatty acid oxidation defects, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 31301 |
| Resource Beneprotein Instant Protein Powder | Carbohydrate disorders | 99557 |

Inherited Metabolic Diseases (IMD) Program
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| OTHER SUPPLEMENTS | | |
|-------------------|---|-----------------|
| Product | Disorder(s) | PIN/ SFS SKU |
| SolCarb | Abetalipoproteinemia, Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | SFS1316 |
| Vitapro | Carbohydrate disorders | SFS0550 |

Inherited Metabolic Diseases (IMD) Program
List of Disorders, Covered Drugs, Supplements and Specialty Foods

Infant Feeds

| Product | Disorder(s) | PIN/ SFS SKU |
|------------------------|---|-----------------|
| Calcilo XD | Hypercalcemia secondary to a listed disease | 91595 |
| Monogen | Fatty Acid Oxidation Defects | 47060 |
| Lipistart | Fatty Acid Oxidation Defects | 47068 |
| Nutramigen A+ | Amino Acid Disorders, Biopterin deficiency, Creatine synthesis/transport deficiency, Glutaric Aciduria II (GA II), Organic Acid Disorders, Pyridoxine (B6) dependent seizures, Urea Cycle Disorders | SFS000046 |
| Portagen Powder | Fatty Acid Oxidation Defects, Mitochondrial disorders | 39581 |
| Pregestimil A+ Powder | Amino acid disorders, Carbohydrate Disorders, Organic acid disorders, Urea cycle disorders | 000045 |
| Ross Carbohydrate Free | Lactic acidosis | 1585 |

Inherited Metabolic Diseases (IMD) Program
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Complete Enteral Feeds

Eligibility criteria for enteral feeds:

Nutrition products will be reimbursed under the IMD program for registered persons when prescribed by a practitioner and when one of the following criteria is met:

- the product is used as part of a treatment plan in addition to medical foods (i.e., modified L-amino acid mixtures) or special low protein foods; OR
- the product is used as part of a treatment plan without medical foods (i.e., modified L-amino acid mixtures) or special low protein foods, and is the primary source of nutrition (e.g., greater than 90% of caloric intake); OR
- the product is used as part of a treatment plan for prevention of hypoglycemia in glycogen storage disorders (continuous and/or bolus feeds)

Exclusion criteria for enteral feeds:

A nutrition product will not be reimbursed under the IMD program if the patient qualifies for reimbursement under the Ontario Drug Benefit (ODB) program or if it is intended for one of the following uses:

- voluntary meal replacement
- convenience
- food allergies
- body building

If the following products are the sole source of nutrition, coverage may be available for eligible persons under the Ontario Drug Benefit Program

| COMPLETE ENTERAL FEEDS | | |
|------------------------------------|------------------|-------------|
| Product | Manufacturer | PIN/SFS SKU |
| Boost 1.0 Standard Chocolate | Nestle Nutrition | 995151 |
| Boost 1.0 Standard Strawberry | Nestle Nutrition | 995153 |
| Boost 1.0 Standard Vanilla | Nestle Nutrition | 995152 |
| Boost 1.5 Plus Calories Chocolate | Nestle Nutrition | 995201 |
| Boost 1.5 Plus Calories Strawberry | Nestle Nutrition | 995202 |
| Boost 1.5 Plus Calories Vanilla | Nestle Nutrition | 995203 |
| Ensure Chocolate | Abbott Nutrition | 914251 |
| Ensure Strawberry | Abbott Nutrition | 914252 |
| Ensure Vanilla | Abbott Nutrition | 914253 |
| Ensure Plus Chocolate | Abbott Nutrition | 914351 |

Inherited Metabolic Diseases (IMD) Program
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| COMPLETE ENTERAL FEEDS | | |
|-----------------------------------|------------------|-------------|
| Product | Manufacturer | PIN/SFS SKU |
| Ensure Plus Strawberry | Abbott Nutrition | 914352 |
| Ensure Plus Vanilla | Abbott Nutrition | 914353 |
| Isosource HN | Nestle Nutrition | 99458 |
| Isosource HN with Fibre | Nestle Nutrition | 99459 |
| Jevity 1 | Abbott Nutrition | 1499 |
| Nutren Junior | Nestle Nutrition | 111912 |
| Nutren Junior Fibre with Prebio | Nestle Nutrition | 111911 |
| Osmolite 1 | Abbott Nutrition | 1497 |
| Pediasure Vanilla | Abbott Nutrition | 91423 |
| Pediasure Chocolate | Abbott Nutrition | 914231 |
| Pediasure Strawberry | Abbott Nutrition | 914232 |
| Pediasure Vanilla with Fibre | Abbott Nutrition | 91424 |
| Pediasure Plus Vanilla with Fibre | Abbott Nutrition | 91422 |
| Peptamen Junior Vanilla | Nestle Nutrition | 11120 |
| Peptamen Unflavoured | Nestle Nutrition | 11100 |
| Resource Kids Essential 1.5 | Nestle Nutrition | 99538 |
| Suplena | Abbott Nutrition | 1441 |
| Tolerex | Nestle Nutrition | 50524 |
| Vital HN | Abbott Nutrition | 1521 |
| Vivonex Pediatric | Nestle Nutrition | SFS0212 |
| Vivonex Plus | Nestle Nutrition | 50545 |

